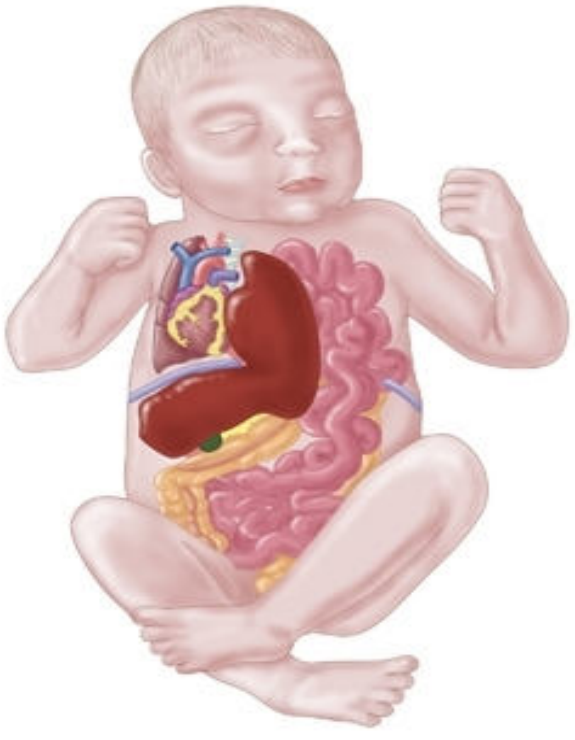
CONGENITAL DIAPHRAGMATIC HERNIA (CDH)



History

- 1679 – Riverius recorded the first CDH
- 1761 – Morgagni desribed types of CDH
- 1905 – Heidenhain repair CDH
- 1925 – Hedbolm suggested that CDH led to pulmonary hypoplasia and early operation improve survival
- 1946 – Gross correct CDH < 24 hours of age
- 1980-1990 – delayed correction become widely accepted

Introduction

- 1 in 2000- 5000 births
- 95.8% posterolateral defects (Bochdalek)
  - 84.4% left side
  - 13.2% right side
  - 2.4% bilateral
- Morgagni and pars sternalis hernias rare
- 10-50% will have associated anomalies.

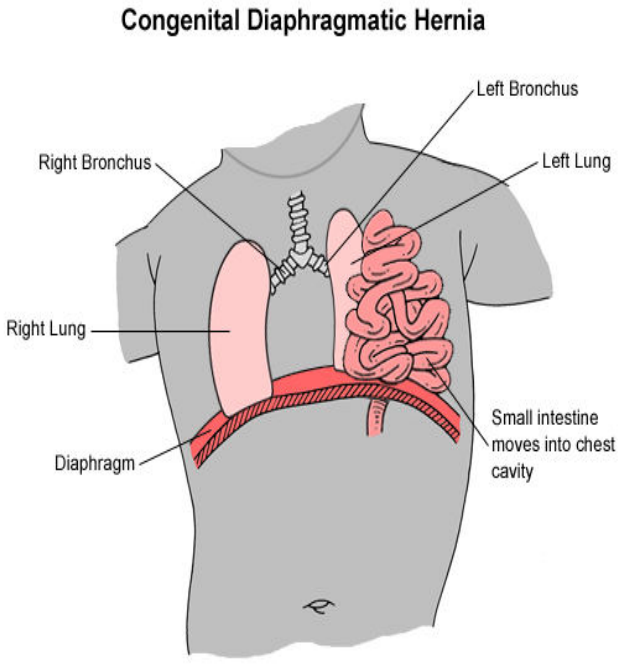
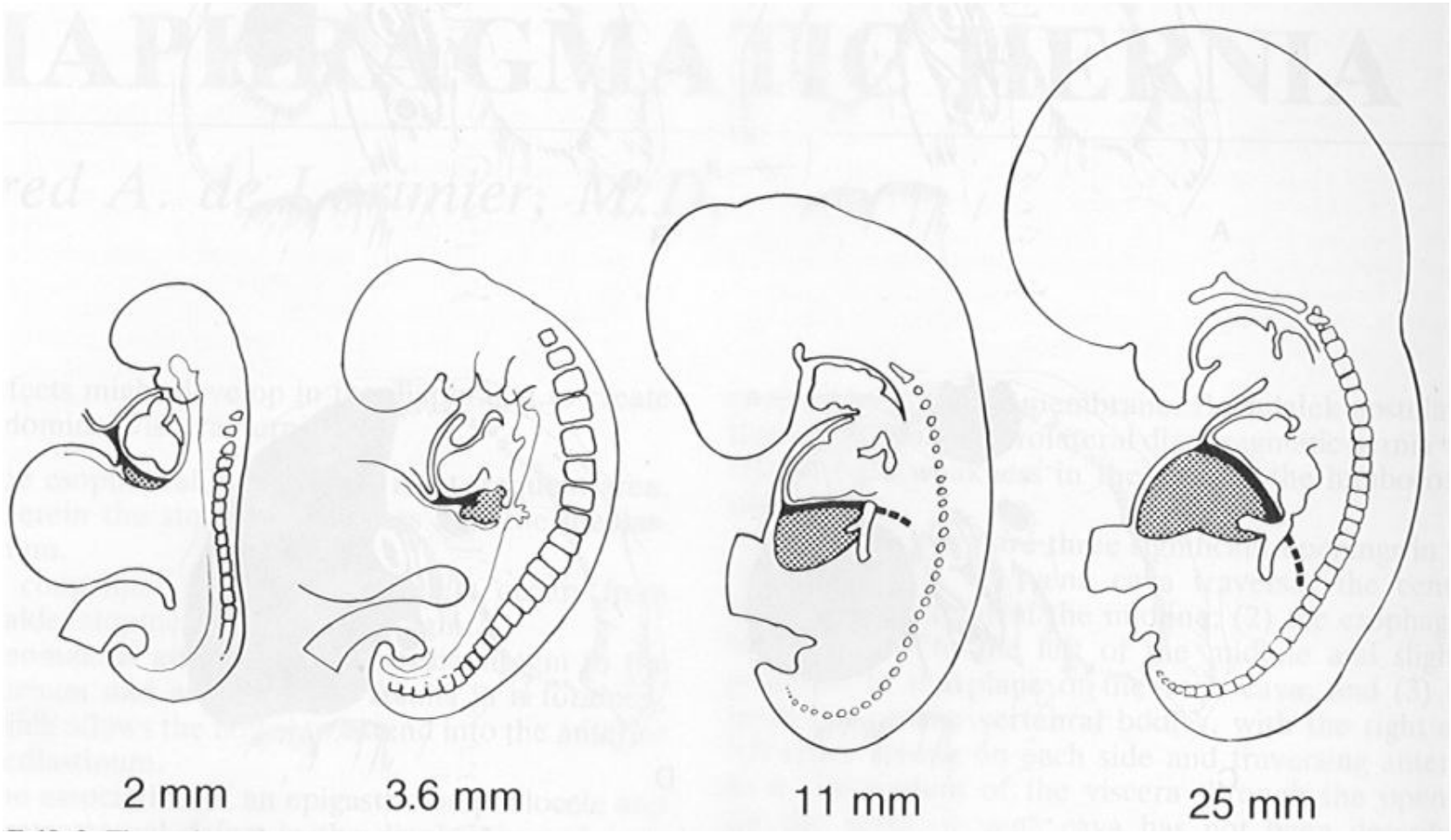


TABLE 1. Organ systems involved in infants with congenital diaphragmatic hernia who have associated malformations

Organ system	Most common major malformations	CDH cases with malformation (%)
Cardiovascular	Ventricular septal defect Hypoplastic heart	36
Genitourinary	Hydronephrosis Polycystic kidneys	23
Central nervous system	Spina bifida Hydrocephalus	15
Skeletal	Hemivertebrate Absent ribs	14
Craniofacial	Cleft palate Facial dysmorphia	7
Gastrointestinal tract and abdominal wall	Gastroschisis Meckel's diverticulum	7

Embryology



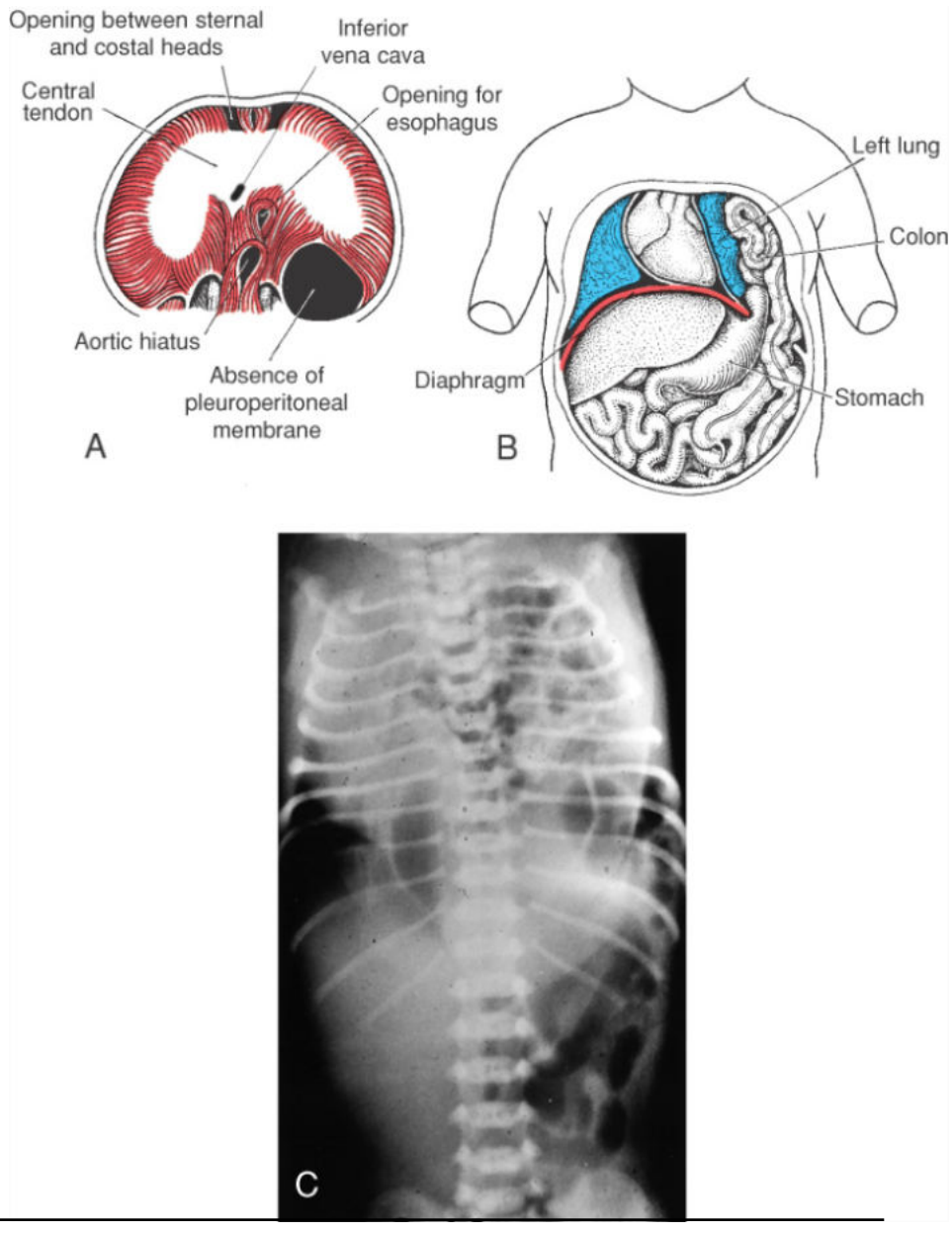
ANATOMY

Diaphragmatic development- begins 4th week

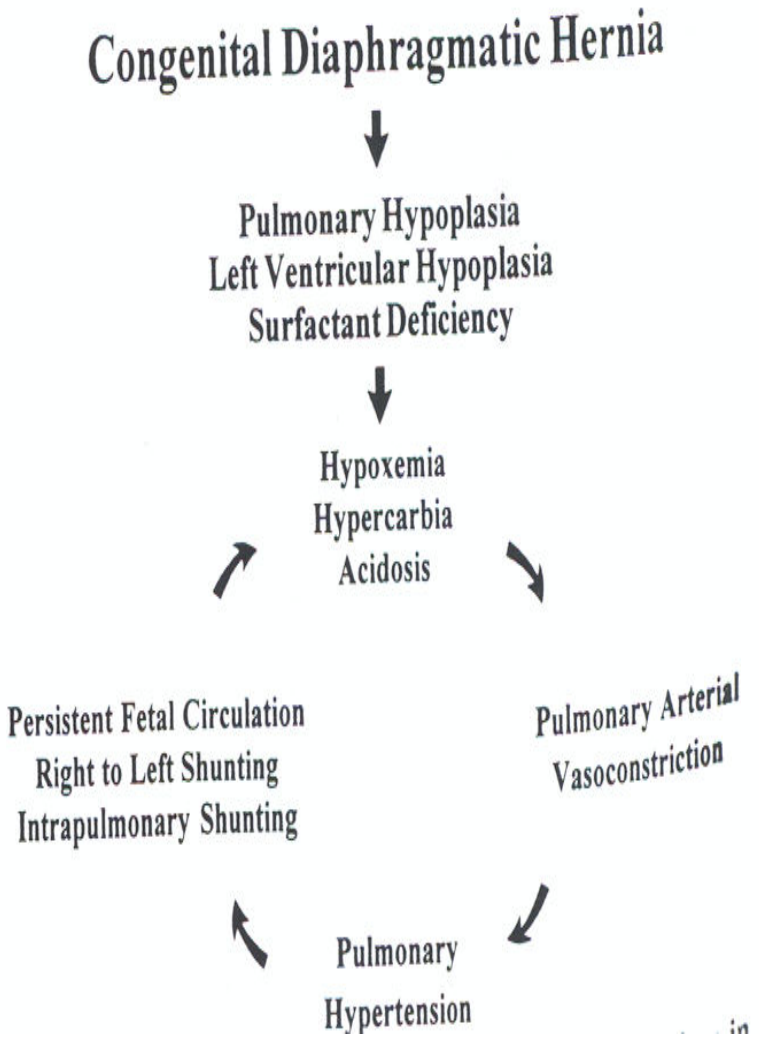
1) Anterior central tendon --Septum transversum (4 wk)- forms the inferior portion of pericardial cavity and defines rudimentary pleuroperitoneal membrane.
2) Dorsolateral portion--Closure of pleuroperitoneal canals and formation of pleuroperitoneal membrane (6-8 wk).
3) Dorsal crura– dorsal esophageal mesentery
4) Muscularisation of diaphragm- from thoracic intercostal muscle group and post hepatic mesenchyme.

Failure of closure of pleuroperitoneal canal (9-10 wk)

Posteriolateral lumbocostal trigone (weak area)- by Bochdalek (1848)



Pathophysiology



- Combination of events…
  A. Physical compression

  B. Pulmonary hypoplasia bilaterally (ipsilateral >contralateral)- Uncorrectable
     A. Smaller bronchi
     B. Less branching
     C. Decreased alveolar SA

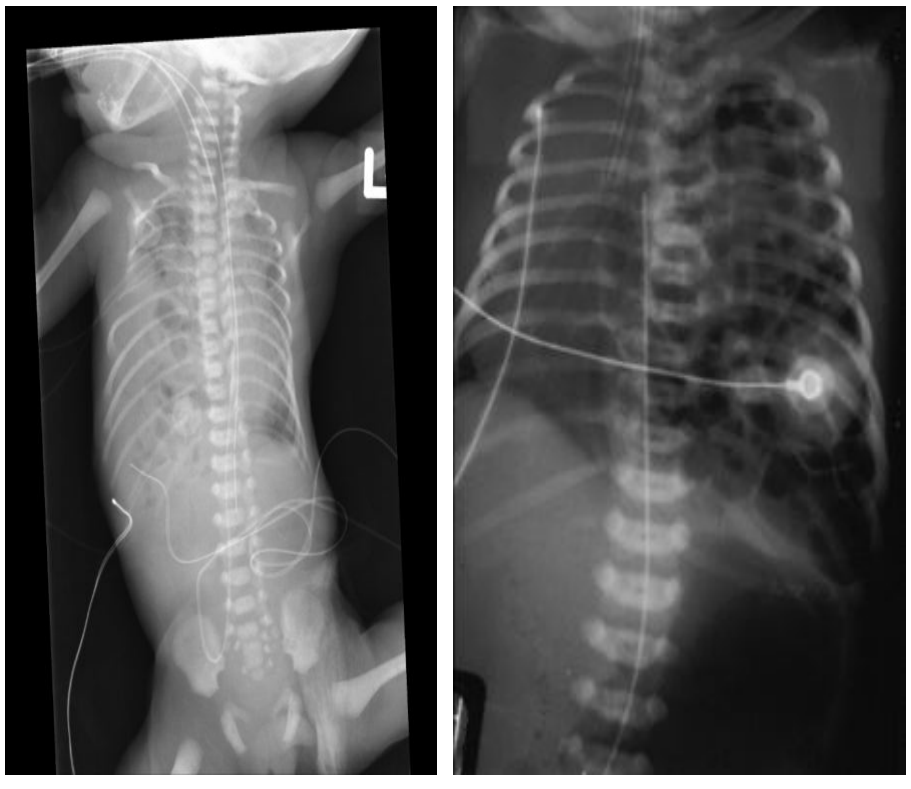
  C. Pulmonary hypertension- potentially reversible
     A. Lack of pulmonary arterioles
     B. Increased thickness of muscle
     - Abnormally high vascular reactivity of vessels
     - Right to left shunt

Diagnosis

- Prenatally
  - U/S – can diagnose 50%, 50% “normal”,
  - as early as 15 weeks
    - » Polyhydramnios
    - » Absent or intrathoracic stomach bubble
    - » Mediastinal or cardiac shift away from hernia
    - » Assess other organs
  - Amniocentesis for chromosomes, alpha fetoprotein (18 weeks)
  - Fetal MRI

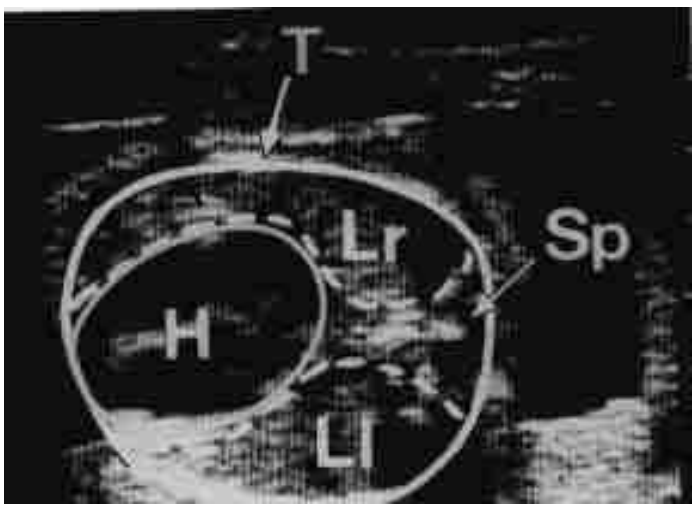
Diagnosis

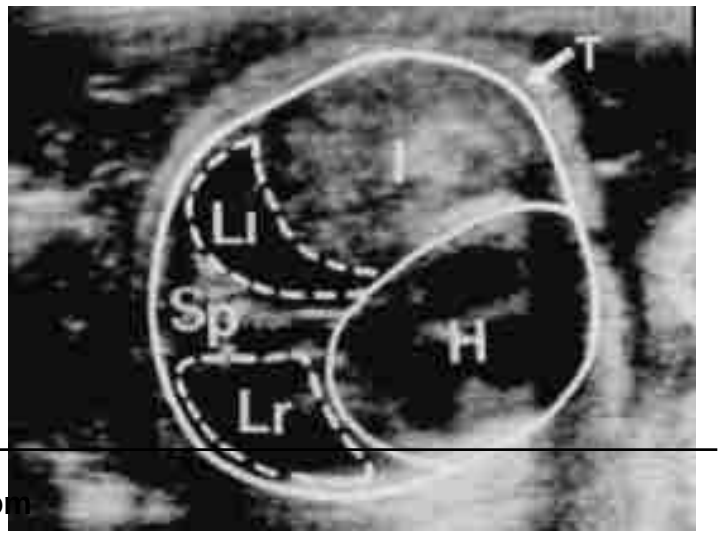
- Postnatal
  - RD in the first 24hours. scaphoid abdomen (majority)
  - After 24h with vomiting, cough, cyanosis, gastric outlet obstruction, herniation of liver, spleen, bowel.



Prognostic Indicators

- *Herniated organs (liver)
- *Lung to head ratio (LHR)
  - <1 poor and <0.85 severe, 0.6-1.35 61% survive, >1.35 100% survive
- Associated anomalies
- Birth weight and Apgar score (the CDH study group)
- Measurement of L/T ratio
  L/T normal ~ 0.52, severe < 0.26
- **Others**- Antenatal diagnosis<24wks, polyhydramnios, position of stomach, right sided defect, Mcgoon's index, ~~Pulm. Artery Index, OI, MVI~~





Prenatal Diagnosis

- Check for associated anomalies
- Plan for delivery at tertiary perinatal centre +/- ECMO
- Options for parents
  - Prenatal therapies, termination, treatment after delivery

Investigations

- Arterial blood gas
  - frequent arterial blood gas (ABG) measurements to assess for pH, PaCO2, and PaO2.
  - PaO2 may be higher from a preductal (right-hand) sampling site.
- Chromosome studies
  - frequent association with chromosomal anomalies (trisomy 13,18)
  - If dysmorphic features are observed on examination, a consultation with a geneticist .
- Serum electrolytes: Monitor serum electrolytes, ionized calcium, and glucose levels initially and frequently. Maintenance of reference range glucose levels and calcium homeostasis is particularly important
- ECHO
- Renal USG

❖ **Pulse oximetry**

- Continuous pulse oximetry is valuable in the diagnosis and management of PPHN.
- Place oximeter probes at preductal (right-hand) and postductal (either foot) sites to assess for a right-to-left shunt at the level of the ductus arteriosus

Management

- Spontaneous Vaginal Delivery, close to term
- Initial resuscitation
  - Supplemental O2
  - No bag & mask
  - ETT with mechanical ventilation
    - Ventilation strategies*
    - Watch for pneumothorax (usually contralateral)
  - NG tube
  - Fluid status
  - Inotropic support
  - Delayed surgery (until patient is stable).

Management

- Mechanical Ventilation
  - May require high setting, high FiO_2
  - High OI (poor survival) ($OI = MAP \times FiO_2 \times 100 / PaO_2$)
- Paralysis/ sedation
- Arterial and venous access
- IV antibiotics, IV fluids, correct acidosis, glucose maintainence
- NG tube
- Inotropic support
- Temperature maintainence
- Role of Tolazoline(a- receptor blocker),CCB, endothelin receptor blockers, Sildenafil, Nitric Oxide
- Role of surfactant- ??

Ventilation Strategies

- Prevent conditions that raise pulmonary vascular resistance (hypoxemia, acidosis, hypotension and hypercarbia) → hyperventilation to control hypoxemia

  Aim: pH > 7.45, PCO_2 < 35 , PaO2 > 60

  → Avoid barotrauma.

Wung et al. showed that some of the mortality in CDH infants was infact due to ventilator-induced lung injury

- Gentle ventilation with permissive hyper-capnia
- Low PIP, pressure-limited ventilation, minimal set RR, minimal sedation and tolerance of high PCO_2 upto 60 → survival>90
- Goal- Preductal PO2> 60mmHg and SaO2> 90% and PCO2< 60mmHg

Surgical Repair

- First repair in 1940 (Ladd & Gross)
- 1970's – Early intervention
  - Emergent surgery to alleviate the compression of the "good" lung
- Current practice allows a period of stabilization prior to repair
  - CDH is a physiologic emergency, not a surgical emergency
  - Pulmonary hypertension is the primary determinant of mortality
  - Wait until weaning / low ventilatory settings or Pulm. HTN abated or stabilised on ECMO

Surgical Repair

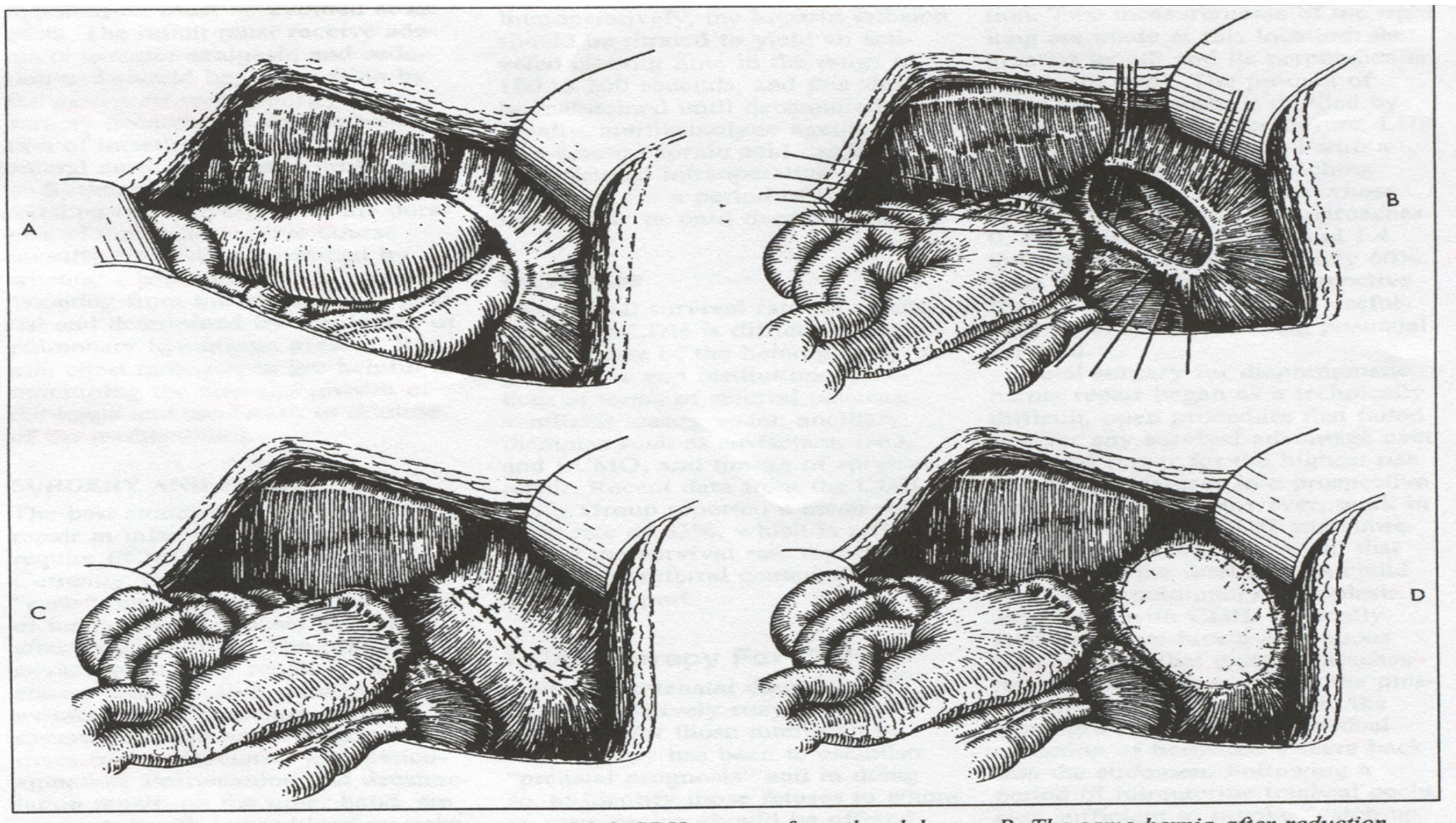


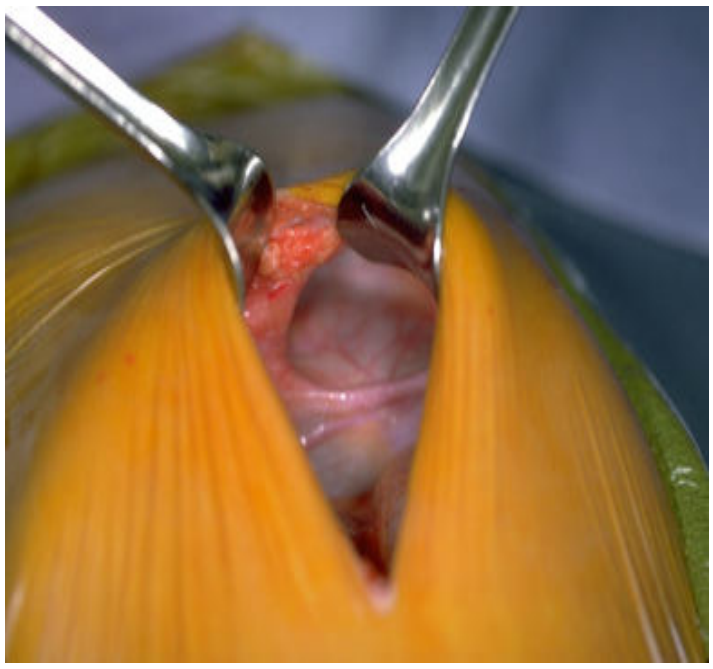
FIGURE 4. A. Schematic drawing of an unreduced left CDH as seen from the abdomen. B. The same hernia after reduction, demonstrating that the spleen is the last organ to be reduced from the chest cavity. Sutures have been placed for a primary repair. C. Completed primary repair of a left CDH. D. Repaired left CDH using prosthetic material. From Spitz L, Coran AG, eds. Rob and Smith's Pediatric Surgery. *London, United Kingdom: Chapman & Hall; 1996.*

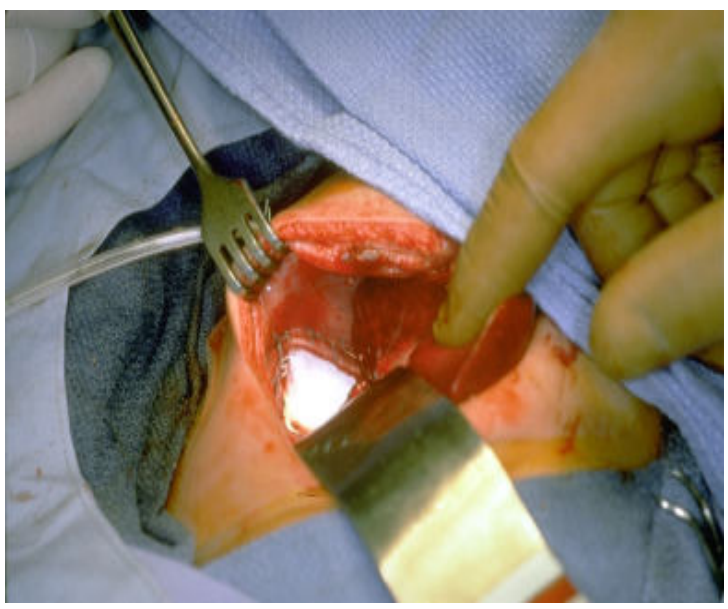
Surgical repair- options

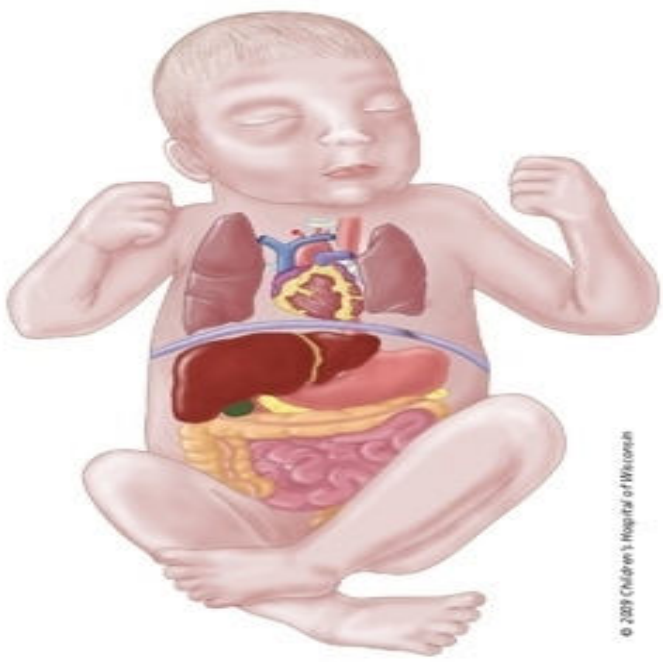
- Open (subcostal incision) / thoracoscopic repair
- Abdominal/ thoracic
- Primary repair after reduction of contents with interrupted non absorbable sutures (Prolene)
- Reconstructive options:

Pre renal fascia, surrounding ribs, Prosthetic patch, split abd. Muscle flap/ LD flap (thoracic)

- Skin closure and creation of ventral hernia/ temporary silo
- ICD – not required (only when ~~bleeding/collection/air leak~~)







Long term outcome

- GERD
- Neurological outcome- CNS and Ear
- Chronic lung disease (obst./rest. Airway- 50% survivors)
- Growth issues
- Chest wall deformities, scoliosis

Newer Advances

1) In utero

– Surgical repair - >70% mortality

– Tracheal ligation or occlusion (FETO)

- 27-28 weeks GA
- Causes distention and secondary hyperplasia
- Good results in 40% of patients

2)Ventilation strategies – HFOV, NO, ECMO, Liquid ventilation (PFC)

3) Lung transplantation

4) Growth factors- combined TRH & steroids

MCQ’s

1) In CDH, the most common defect is

a. Anterior morgagni
b. Posteriolateral Bochdalek
c. Right sided defect
d. Pars sternalis

2) CDH is a/an

a. Physiologic/ medical emergency
b. Surgical emergency
c. Both a and b
d. OPD case

3) 2kg newborn child Day 1, with respiratory distress, Scaphoid abdomen, Bowel sounds +nt in left chest and X-ray chest s/o bowel loops present in left chest with mediastinal shift to right. The first intervention you do in emergency

a. Bag and mask
b. Iv Fluids and antibiotics
c. ETT and ventilation
d. Ng tube insertion

4) Antenatal Usg s/o Polyhydramnios and absent stomach bubble, most prabable diagnosis s/o

a. PUV
b. Malrotation
c. CDH
d. TEF with EA

5) Day 4 Child with CDH on ventilator with high settings, ECHO s/o PDA, referred from NiCU for surgical opinion

a. Immediate surgery
b. Wait further
c. ECMO
d. PDA ligation

6) Child with CDH on ventilator requires

a. Aggressive ventilation with high PIP
b. Gentle ventilation with permissive hypercapnia
c. High RR
d. High tidal volume

7) CDH associated with better prognosis is

a. Liver up
b. Premature
c. LHR > 1
d. Right sided

8) Antenatal review to pediatric surgery OPD, G1P1,30wks pregnant with CDH child with polyhydramnios

a. Termination of pregnancy
b. Prenatal steroids and delivery at 34 wks
c. SVD near term at hospital
d. Refer to genecist

9) Surgery for CDH

a. Reduction of contents and primary repair
b. Primary repair after content reduction with ventral hernia creation
c. Prosthetic Patch repair
d. Thoracoscopic repair
e. All of the above

10) Recent advances in treatment of CDH antenatally

a. ECMO
b. HFOV
c. FETO
d. NO